Complete Summary

GUIDELINE TITLE

Pulmonary artery hypertension and sleep-disordered breathing: ACCP evidence-based clinical practice guidelines.

BIBLIOGRAPHIC SOURCE(S)

Atwood CW Jr, McCrory D, Garcia JG, Abman SH, Ahearn GS. Pulmonary artery hypertension and sleep-disordered breathing: ACCP evidence-based clinical practice guidelines. Chest 2004 Jul;126(1 Suppl):72S-77S. [27 references] PubMed

GUIDELINE STATUS

This is the current release of the guideline.

COMPLETE SUMMARY CONTENT

SCOPE

DISEASE/CONDITION(S)

- Pulmonary arterial hypertension
- Sleep-disordered breathing (includes central and obstructive sleep apnea and nocturnal desaturation)

GUIDELINE CATEGORY

Evaluation
Management
Treatment

CLINICAL SPECIALTY

Cardiology
Internal Medicine
Pulmonary Medicine
Sleep Medicine

INTENDED USERS

Physicians

GUIDELINE OBJECTIVE(S)

To review the available data on the relationship between sleep-disordered breathing (SDB) and pulmonary arterial hypertension (PAH), with a focus on the prevalence of SDB in patients with idiopathic PAH (IPAH); the prevalence of PAH in patients with SDB; and the effects of SDB treatment on PAH

TARGET POPULATION

Patients with pulmonary arterial hypertension and sleep-disordered breathing

INTERVENTIONS AND PRACTICES CONSIDERED

Evaluation/Management/Treatment

1. Assessment of sleep-disordered breathing in patients with pulmonary arterial hypertension
2. Polysomnography
3. Routine evaluation for the presence of pulmonary arterial hypertension in patients with obstructive sleep apnea
4. Positive airway pressure therapy

MAJOR OUTCOMES CONSIDERED

- Prevalence of sleep-disordered breathing in patients with pulmonary arterial hypertension
- Prevalence of pulmonary arterial hypertension in patients with sleep apnea
- Effect of sleep apnea therapy on pulmonary hemodynamics in patients with pulmonary arterial hypertension

METHODOLOGY

METHODS USED TO COLLECT/SELECT EVIDENCE

Hand-searches of Published Literature (Primary Sources)
Hand-searches of Published Literature (Secondary Sources)
Searches of Electronic Databases

DESCRIPTION OF METHODS USED TO COLLECT/SELECT THE EVIDENCE

Note from National Guideline Clearinghouse (NGC): The Center for Clinical Health Policy Research at Duke University identified and evaluated evidence on this topic, working with the guideline development panel to formulate key questions suitable for systematic literature synthesis.

Search Strategy

The American College of Chest Physicians Committee on Clinical Practice Guidelines for Pulmonary Arterial Hypertension formulated three key questions to be answered by a comprehensive critical review of the published evidence regarding sleep-disordered breathing (SDB) and pulmonary arterial hypertension (PAH): (1) what is the prevalence of SDB among patients with idiopathic PAH (IPAH), (2) what is the prevalence of PAH among patients with SDB, and (3) does treatment of obstructive sleep apnea (OSA) affect pulmonary hemodynamics in patients with OSA and PAH? To address these questions, the guideline committee conducted a computerized search of the MEDLINE bibliographic database from 1992 to October 2002. The guideline committee searched using the term *hypertension*, *pulmonary* combined with *sleep apnea syndromes* and subheadings, and *sleep apnea*, *obstructive* and subheadings. The search was limited to articles concerning human subjects that were published in English and accompanied by an abstract. In addition, the guideline committee searched the reference lists of included studies, practice guidelines, systematic reviews, and meta-analyses, and consulted with clinical experts to identify relevant studies undetected by the search strategy or published before 1992.

Study Selection

The guideline committee considered studies conducted among patients with known or suspected IPAH, as well as populations with known or suspected SDB. The committee excluded studies of neonates and studies of patients with chronic obstructive pulmonary disease (COPD) or coronary artery disease. They accepted polysomnography or four-channel cardiopulmonary sleep studies to ascertain the presence or absence of SDB. The committee accepted right-heart catheterization or echocardiography for the diagnosis of PAH and the evaluation of hemodynamic response to treatment. They accepted continuous positive airway pressure (CPAP) or surgical intervention as recognized treatments for OSA. They excluded case series with <10 subjects.

Two physicians, one with methodologic expertise and one with content area expertise, reviewed the abstracts of candidate articles and selected a subset for review in full text. Full-text articles were reviewed by both physicians to determine whether they were original investigations or review articles and whether they were pertinent to at least one of the key questions.

NUMBER OF SOURCE DOCUMENTS

Not stated

METHODS USED TO ASSESS THE QUALITY AND STRENGTH OF THE EVIDENCE

Expert Consensus
Weighting According to a Rating Scheme (Scheme Given)

RATING SCHEME FOR THE STRENGTH OF THE EVIDENCE

Quality of the Evidence

Good = evidence based on good randomized controlled trials or meta-analyses

Fair = evidence based on other controlled trials or randomized controlled trials with minor flaws

Low = evidence based on nonrandomized, case-control, or other observational studies

Expert opinion = evidence based on the consensus of the carefully selected panel of experts in the topic field. There are no studies that meet the criteria for inclusion in the literature review.

METHODS USED TO ANALYZE THE EVIDENCE

Systematic Review with Evidence Tables

DESCRIPTION OF THE METHODS USED TO ANALYZE THE EVIDENCE

Not stated

METHODS USED TO FORMULATE THE RECOMMENDATIONS

Informal Consensus

DESCRIPTION OF METHODS USED TO FORMULATE THE RECOMMENDATIONS

An international panel of 19 experts representing five medical experts was assembled. Representatives from other medical and patient advocacy associations were also invited to join the panel (including the American College of Cardiology, American College of Rheumatology, and the Pulmonary Hypertension Association). These experts convened on several occasions, including the culminating panel conference in September 2003, in which they deliberated over the composition of the final recommendations and grading of the current state of the evidence, benefits to the patient, and the strength of the recommendations.

Guideline development was led by an executive committee including the chair, the leader of the methodology support group, and the American College of Chest Physicians project manager, which supervised the guideline development process, methodologic issues, panel composition, structure of the final document, and activities of the writing committees. Each writing committee, led by a group leader who served as primary author and editor of that chapter, conferred with the methodology team on inclusion/exclusion criteria, relevant research questions, and important literature that was not readily identified. These individuals continue

with their responsibilities to assist in the development of the implementation tools.

When the evidence was insufficient for evidence-based recommendations, the panel used informal group consensus techniques to develop recommendations based on the expert opinion of the panel. With every member of the panel attending the final conference, the expert-based opinions are truly representative of geographically diverse and multispecialty inclusive practice patterns of the complete panel.

RATING SCHEME FOR THE STRENGTH OF THE RECOMMENDATIONS

Strength of Recommendations

A = strong recommendation
B = moderate recommendation
C = weak recommendation
D = negative recommendation
I = no recommendation possible (inconclusive)
E/A = strong recommendation based on expert opinion only
E/B = moderate recommendation based on expert opinion only
E/C = weak recommendation based on expert opinion only
E/D = negative recommendation based on expert opinion only

Net Benefit

Substantial
Intermediate
Small/weak
None
Conflicting
Negative

COST ANALYSIS

A formal cost analysis was not performed and published cost analyses were not reviewed.

METHOD OF GUIDELINE VALIDATION

External Peer Review
Internal Peer Review

DESCRIPTION OF METHOD OF GUIDELINE VALIDATION

The writing groups and the executive committee of the panel extensively reviewed each chapter during the writing process. The final conference provided an opportunity for the entire panel to review the latest drafts. Following final revisions and one final review by the executive committee, each chapter of the guidelines was reviewed and approved by the American College of Chest Physicians (ACCP) Health and Science Policy Committee, the ACCP Pulmonary

Vascular NetWork, and then by the ACCP Board of Regents. The guidelines have not been field tested.

RECOMMENDATIONS

MAJOR RECOMMENDATIONS

Rating schemes for level of evidence, strength of recommendation, and net benefit follow the "Major Recommendations."

Based on the available evidence, the following recommendations were accepted by the American College of Chest Physicians Guidelines committee for the evaluation of sleep disordered breathing in the setting of idiopathic pulmonary arterial hypertension (IPAH), for the evaluation pulmonary arterial hypertension (PAH) in the setting of obstructive sleep apnea (OSA), and for the treatment of OSA and its effect on PAH.

1. In the evaluation of patients with pulmonary arterial hypertension (PAH), an assessment of sleep-disordered breathing (SDB) is recommended. **Quality of evidence: low; net benefit: small/weak; strength of recommendation: C**.
2. In the evaluation of a patient with PAH for SDB, polysomnography is recommended if obstructive sleep apnea (OSA) is suspected as the etiology, if a screening test result for OSA is positive, or if a high clinical suspicion for OSA is present. **Quality of evidence: expert opinion; net benefit: intermediate; strength of recommendation: E/B**.
3. In the management of patients with OSA, routine evaluation for the presence of PAH is not recommended. **Quality of evidence: low; net benefit: none; strength of recommendation: I**.
4. In patients with OSA and PAH, treatment of OSA with positive airway pressure therapy should be provided with the expectation that pulmonary pressures will decrease, although they may not normalize, particularly when PAH is more severe. **Quality of evidence: low; net benefit: small/weak; strength of recommendation: C**.

Definitions

Quality of the Evidence

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Low = evidence based on nonrandomized, case-control, or other observational studies

Expert opinion = evidence based on the consensus of the carefully selected panel of experts in the topic field. There are no studies that meet the criteria for inclusion in the literature review.

Strength of Recommendations

A = strong recommendation
B = moderate recommendation
C = weak recommendation
D = negative recommendation
I = no recommendation possible (inconclusive)
E/A = strong recommendation based on expert opinion only
E/B = moderate recommendation based on expert opinion only
E/C = weak recommendation based on expert opinion only
E/D = negative recommendation based on expert opinion only

Net Benefit

Substantial
Intermediate
Small/weak
None
Conflicting
Negative

CLINICAL ALGORITHM(S)

None provided

EVIDENCE SUPPORTING THE RECOMMENDATIONS

TYPE OF EVIDENCE SUPPORTING THE RECOMMENDATIONS

The type of supporting evidence is identified and graded for each recommendation (see "Major Recommendations").

BENEFITS/HARMS OF IMPLEMENTING THE GUIDELINE RECOMMENDATIONS

POTENTIAL BENEFITS

Based on the limited data available, the guideline committee concluded that the severity of the pulmonary arterial hypertension (PAH) in patients with obstructive sleep apnea (OSA) is mild, and that continuous positive airway pressure (CPAP) therapy is moderately effective in reducing pulmonary arterial pressure in this setting. No data have been reported on functional improvement, quality of life, or other patient-level outcomes in patients with OSA and PAH.

POTENTIAL HARMS

Not stated

QUALIFYING STATEMENTS

QUALIFYING STATEMENTS

- The information provided in the guideline should be used in conjunction with clinical judgment. Although the guideline provides recommendations that are based on evidence from studies involving various populations, the recommendations may not apply to every individual patient. It is important for the physician to take into consideration the role of patient preferences and the availability of local resources.
- The American College of Chest Physicians (ACCP) is sensitive to concerns that nationally and/or internationally developed guidelines are not always applicable in local settings. Further, guideline recommendations are just that, recommendations not dictates. In treating patients, individual circumstances, preferences, and resources do play a role in the course of treatment at every decision level. Although the science behind evidence-based medicine is rigorous, there are always exceptions. The recommendations are intended to guide healthcare decisions. These recommendations can be adapted to be applicable at various levels.

IMPLEMENTATION OF THE GUIDELINE

DESCRIPTION OF IMPLEMENTATION STRATEGY

Implementation tools are being developed, including a quick reference guide in print and personal digital assistant format, and educational slide presentations for physicians and other health-care practitioners.

INSTITUTE OF MEDICINE (IOM) NATIONAL HEALTHCARE QUALITY REPORT CATEGORIES

IOM CARE NEED

Living with Illness

IOM DOMAIN

Effectiveness

IDENTIFYING INFORMATION AND AVAILABILITY

BIBLIOGRAPHIC SOURCE(S)

Atwood CW Jr, McCrory D, Garcia JG, Abman SH, Ahearn GS. Pulmonary artery hypertension and sleep-disordered breathing: ACCP evidence-based clinical practice guidelines. Chest 2004 Jul;126(1 Suppl):72S-77S. [27 references] PubMed

ADAPTATION

Not applicable: The guideline was not adapted from another source.

DATE RELEASED

2004 Jul

GUIDELINE DEVELOPER(S)

American College of Chest Physicians - Medical Specialty Society

SOURCE(S) OF FUNDING

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GUIDELINE COMMITTEE

American College of Chest Physicians (ACCP) Expert Panel on Pulmonary Artery Hypertension

COMPOSITION OF GROUP THAT AUTHORED THE GUIDELINE

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FINANCIAL DISCLOSURES/CONFLICTS OF INTEREST

The following participants have disclosed information regarding potential or real conflicts of interest and commitment:

Steven H. Abman, MD: scientific advisory board for INO Therapeutics; consultant for Pfizer.

Charles W. Atwood, Jr., MD, FCCP: research support from Respironics, Inc.

David B. Badesch, MD, FCCP: consultant or Speaker's Bureau for Glaxo Wellcome/GlaxoSmithKline, Actelion, InterMune, Encysive, Myogen, Astra-Merck, Astra-Zeneca, Exhale Therapeutics/CoTherix, Forrest Labs, INO Therapeutics, Berlex; research support from Glaxo Wellcome/GlaxoSmithKline, United Therapeutics, Boehringer Ingelheim, Actelion, Encysive, ICOS/Texas Biotechnologies/Encysive, Myogen, INO Therapeutics, Scleroderma Foundation, National Institutes of Health, National Heart, Lung, and Blood Institute, United Therapeutics, Pfizer, American Lung Association.

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Ramona L. Doyle, MD, FCCP: Speaker's Bureau for Actelion; clinical research for Actelion, Myogen, United Therapeutics.

David D. Gutterman, MD, FCCP: stock options with Johnson & Johnson; relative who is a Vice-President at GlaxoSmithKline.

James E. Loyd, MD, FCCP: relationships with GlaxoSmithKline, United Therapeutics, Actelion, ICOS/Texas Biotechnology, Westat, PRA International, Pfizer, Exhale Therapeutics.

Michael D. McGoon, MD: past research support from Glaxo Wellcome, United Therapeutics, Actelion; research support from Texas Biotech/Encysive, Myogen, Pfizer, Medtronic.

Vallerie V. McLaughlin, MD, FCCP: consultant for Actelion, United Therapeutics, Exhale Therapeutics; Speaker's Bureau for Actelion; research funding from Actelion, United Therapeutics, Pfizer, Encysive/Texas Biotechnologies, Glaxo Wellcome, Exhale Therapeutics, Myogen.

Stuart Rich, MD: research funding from Actelion, Pfizer, United Therapeutics, Encysive, Myogen; consultant for Actelion, Pfizer, United Therapeutics, GlaxoSmithKline.

Lewis J. Rubin, MD, FCCP: consultant for Actelion, Myogen, Schering, Exhale Therapeutics, United Therapeutics, Pfizer, Celgene; investigator for Actelion, Myogen, Exhale, Pfizer, Celgene; no stock holdings or other ownerships or positions.

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Virginia D. Steen, MD: relationships with Arthritis Foundation, Scleroderma Foundation, Actelion.

Fredrick M. Wigley, MD: research funding from Biogen, Pfizer, Actelion; consultant to Genzyme.

GUIDELINE STATUS

This is the current release of the guideline.

GUIDELINE AVAILABILITY

Electronic copies: Available to subscribers of Chest - The Cardiopulmonary and Critical Care Journal.

Print copies: Available from the American College of Chest Physicians, Products and Registration Division, 3300 Dundee Road, Northbrook IL 60062-2348.

AVAILABILITY OF COMPANION DOCUMENTS

The following are available:

Background Articles

- Rubin, LJ. Diagnosis and management of pulmonary arterial hypertension: ACCP evidence-based clinical practice guidelines. Introduction. Chest 2004 Jul;126(1 Suppl):7S-10S.
- Rubin LJ. Diagnosis and management of pulmonary arterial hypertension: ACCP evidence-based clinical practice guidelines. Executive summary. Chest 2004 Jul;126(1 Suppl):4S-6S.
- McCrory DC, Lewis SZ. Methodology and grading for pulmonary hypertension evidence review and guideline development. Chest 2004 Jul;126(1 Suppl):11S-13S.

Electronic copies: Available to subscribers of Chest - The Cardiopulmonary and Critical Care Journal.

Print copies: Available from the American College of Chest Physicians, Products and Registration Division, 3300 Dundee Road, Northbrook IL 60062-2348.

PATIENT RESOURCES

None available

NGC STATUS

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